A hidden cause of hemoptysis: coronary artery-to-pulmonary parenchyma fistula

Hemoptizinin gizli bir nedeni: Koroner arter ile pulmoner parenkim arasında fistül

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Summary – A 60-year-old man presented with complaints of chronic hemoptysis present for many years and a six-month history of chest pain. Physical examination showed a grade II/VI continuous murmur at the left sternal border. Electrocardiography showed normal sinus rhythm and nonspecific ST-T changes in lateral leads. Echocardiography showed mild left ventricular hypertrophy. Exercise test was discontinued because of anginal symptoms and occurrence of lateral ST depression. Hemoptysis was observed a few times during hospitalization. Computed tomography of the thorax showed no abnormality to explain hemoptysis. Coronary angiography revealed a critical lesion in the left anterior descending artery and a large, tortuous right coronary artery with a fistulization tract originating from its proximal region and draining into the left lung parenchyma. The lesion in the left anterior descending artery was stented and percutaneous coil embolization of the fistula was performed in another session. Coronary angiography showed complete occlusion of the fistula and no residual shunting. In the six-month period after the procedure, the patient was free of symptoms of angina and hemoptysis.

Özet – Altmış yaşında erkek hasta, yıllardır varlığını sürdüren kronik kanlı balgam çıkarma ve altı aydır var olan göğüs ağrısı yakınmalarıyla başvurdu. Fizik muayenede, sol sternal kenarda II/IV şiddetinde sürekli üfürüm duyuldu. Elektrokardiyografide normal sinüs ritmi ve lateral derivasyonlarda spesifik olmayan ST-T değişiklikleri, ekokardiyografide hafif sol ventrikül hipertrofisi izlendi. Egzersiz testi, anginal semptomlar ve lateral ST çökmesi ortaya çıkması üzerine sonlandırıldı. Hastane izlemi sırasında hastanın birkaç kez kanlı balgam çıkardığı görüldü; ancak, göğüs bilgisayarlı tomografisinde bu durumu açıklayacak bir neden bulunamadı. Koroner anjiyografide sol ön inen arterde kritik bir lezyon ve büyük, kıvrımlı sağ koroner arterin proksimalinden köken alan ve sol akciğer parenkimine boşalan fistül kanalı saptandı. Sol ön inen arterdeki kritik lezyona stent yerleştirildi ve başka bir seansta fistüle perkütan koil embolizasyonu uygulandı. İşlemden sonra yapılan koroner anjiyografide fistülün tam tıkandığı ve kaçak olmadığı izlendi. Altı aylık izlem sırasında hastanın göğüs ağrısı ve kanlı balgam çıkarma yakınmaları bir daha görülmedi.

Coronary artery fistula is a rare congenital or acquired anomalous shunt from a coronary artery to a cardiac chamber or great vessel. It is seen in 0.6-1.5% of patients who undergo coronary angiography and in 0.002% of the general population.[1] It is the most common congenital coronary anomaly leading to significant hemodynamic abnormalities.[2] This abnormality results from the persistence of intratrabecular gaps that are normally present in the intrauterine life. Most commonly, CAF arises from the right coronary artery (50%) with drainage mostly to the right ventricle (41%). While most CAFs are small and asymptomatic, they may cause many problems including continuous murmur, angina pectoris, acute myocardial infarction, sudden death, coronary steal, congestive heart failure, infective endocarditis, arrhythmia, coronary aneurysm formation, and superior vena cava syndrome.

Abbreviations:

CAF *Coronary artery fistula*
RCA *Right coronary artery*

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We describe a case of CAF with associated hemoptysis, a condition that has been reported in only two cases.

CASE REPORT

A 60-year-old male patient presented to our hospital with chronic nonmassive hemoptysis present for many years and a six-month history of chest pain. He was a nonsmoker and had previously been diagnosed with hypertension and hyperlipidemia. Physical examination showed a grade II/VI continuous murmur heard at the left sternal border. Blood pressure was normal. Electrocardiography showed normal sinus rhythm with a heart rate of 73 bpm and nonspecific ST-T changes in lateral leads. Telecardiogram showed a normal cardiothoracic index and remarkable pulmonary arteries. Echocardiography showed mild left ventricular hypertrophy and an ejection fraction of 60%. Maximal exercise treadmill test was stopped because of anginal symptoms with lateral ST-segment depression during stage II of the Bruce protocol. Laboratory tests were unremarkable including coagulation, infection, and tuberculosis tests. Hemoptysis was observed a few times during his hospital course. Computed tomography of the thorax showed no major abnormalities to explain hemoptysis. Then, the patient was referred to the catheterization laboratory for coronary angiography. Selective coronary angiography revealed a critical lesion in the left anterior descending artery, noncritical plaques in the circumflex artery, and a large, tortuous RCA with a fistulization tract originating from the proximal RCA, crossing the base of the heart, and draining into the left lung parenchyma (Fig. 1a, b). A 3 x 12-mm bare metal stent (Driver, Medtronic Minneapolis, Minnesota, USA) was placed in the left anterior descending artery without any complications. The CAF was suitable for percutaneous coil embolization, thus we decided to occlude the fistula in another session. Selective injection of contrast media into the RCA via a 6-Fr Judkins right guiding catheter revealed an aberrant and tortuous fistulous tract from the proximal segment of the RCA to the left lung field. A runthrough 0.014-inch guide wire (Terumo, Japan) and a microcatheter were passed through the guiding catheter to the RCA. After the guide wire was removed, a total of six Multi-Loop-18 Fibered Platinum Coils (Boston Scientific, Natick, Massachusetts, USA) were placed with controlled delivery to embolize the CAF. After embolization, selective coronary angiography showed complete occlusion of the fistula and no residual shunting (Fig. 1c). In the six-month period after the procedure, the patient was free of symptoms of angina and hemoptysis.

DISCUSSION

Coronary artery fistula refers to a direct connection between a coronary artery and one of the cardiac chambers, large vessels, or other vascular structures.[3] It is most commonly diagnosed incidentally during coronary angiography.[4] Clinical manifestations vary depending on the site and size of the shunt, pressure gradient between the origin and drainage site, and underlying valvular or coronary pathologies. Most patients are asymptomatic. However, myocardial ischemia, congestive heart failure, arrhythmia, sudden death, infective endocarditis, and rupture may devel-

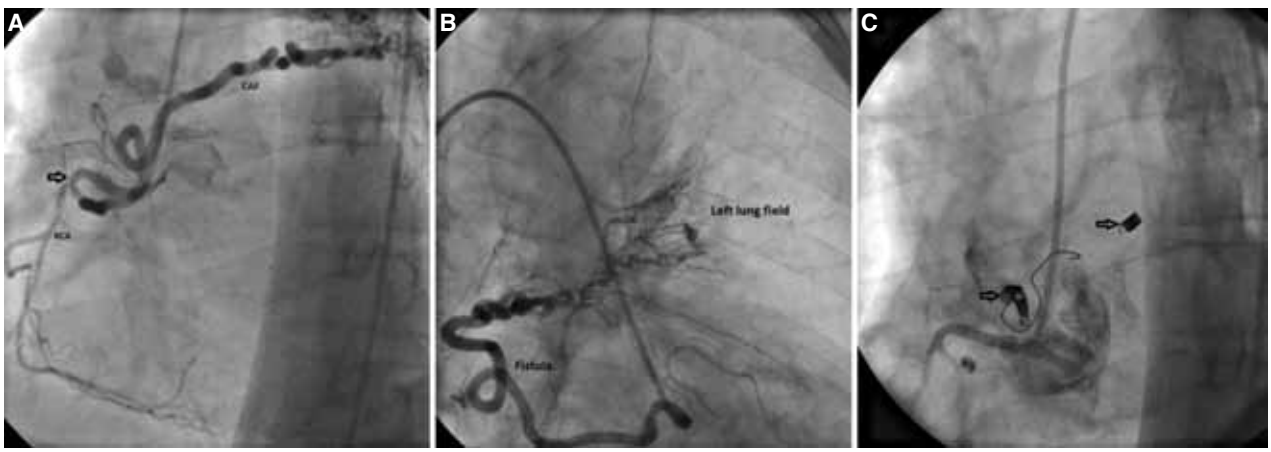

Figure 1. **(A, B)** Coronary artery fistula from the proximal right coronary artery, draining into the left pulmonary parenchyma. **(C)** One 4 x 12-mm and five 3 x 50-mm fibered platinum coils were delivered by a coil pusher and successful occlusion of the flow was achieved.

op.[5] Hemopericardium due to a spontaneously ruptured aneurysmal fistula has also been reported.[6] In rare cases, thrombus may be formed within the fistula and cause myocardial infarction.[7]

The right coronary artery or its branches have been reported as the most frequent artery for fistula origin and low-pressure chambers as the most common regions of drainage. These drainage sites can be listed in descending order of frequency as the right ventricle (41%), right atrium (26%), pulmonary artery (17%), left ventricle (3%), and superior vena cava (1%).[8] A rare coronary fistulization to the right lower pulmonary lobe parenchyma has also been reported.[9] In the present case, the fistula originated from the RCA and the draining site was the left lung parenchyma.

The management of CAF is controversial, particularly in cases of small and asymptomatic fistulas. Coronary fistulas require treatment in the presence of large fistulas, progressive left-to-right shunt, myocardial ischemia, left ventricular dysfunction, congestive heart failure, and in order to prevent endocarditis/endarteritis. There are two interventional treatment modalities for CAF: surgical ligation and percutaneous transcatheter approach, both having similar rates of efficacy, mortality, and morbidity.[10] Transcatheter embolization technique can be used instead of surgery in selected cases. This method may be preferred especially in the absence of a major branch that may accidentally be embolized, in cases where the coronary artery branch supplying the fistula can safely be cannulized, and in a CAF with single fistula origin and drainage. The most important point for catheter-based treatment is to occlude the fistula as distally as possible, closest to its end point. This would preserve the side branches supplying the normal myocardium. In order to reduce complications and increase success rate, the catheter and other equipment should be selected based on the morphological features of the fistula. In our case, catheter-based closure using controlled-release coil embolization resulted in successful occlusion of the CAF without any complications.

In the present case, we thought that the CAF was the cause of hemoptysis because there was no other etiology. In the literature, we found only two reports on the association between hemoptysis and CAF. The first one was a 58-year-old man with hemoptysis and large fistula from the circumflex artery to the pulmonary system.[11] After successful coil embolization of the fistula, junctional bradycardia developed as a complication of small-branch occlusion probably feeding the sinus node. The second was a 6-year-old boy who had tetralogy of Fallot and CAF arising from the RCA to bilateral hilar lung fields.[12] In both cases, hemoptysis disappeared promptly after coil embolization, as in our patient.

In conclusion, there are many symptoms and complications attributable to CAFs. However, to the best of our knowledge, there are few cases to implicate a coronary fistula as the cause of chronic hemoptysis. Physicians should be alert to the possibility of a CAF in a patient with undetermined cause of hemoptysis.

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Key words: Coronary angiography; coronary vessel anomalies; embolization, therapeutic; fistula/therapy; hemoptysis/etiology.

Anahtar sözcükler: Koroner anjiyografi; koroner damar anomalisi; embolizasyon/terapötik; fistül/tedavi; hemoptizi.